I. AMENDMENTS

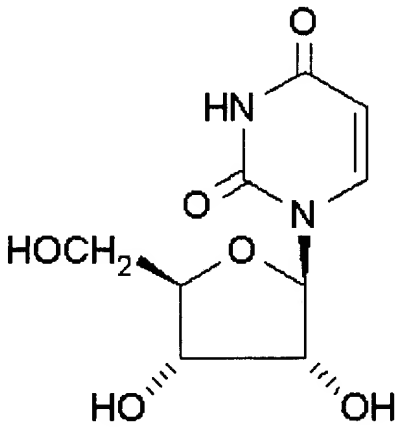
In the Claims

Please cancel claims 1 to 6, 8 to 27 and 66 without prejudice and add new claims 67 to 94.

Upon entry of the present amendment, the claims will stand as follows in the present application:

Claims 1-66 (Canceled).

67. (New) A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder an effective amount of a compound of Formula I:



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68. (New) A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of uridine.

69. (New) A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of 1-b-D-ribofuranosyluracil.

70. (New) The method according to claim 67, wherein the mitochondrial disorder is a primary disorder comprising at least one mutation in mitochondrial or nuclear DNA.

71. (New) The method according to claim 67, wherein the mitochondrial disorder is selected from the group consisting of Huntington's disease, Amyotrophic lateral sclerosis, MELAS (Mitochondrial encephalomyopathy with lactic acidemia and stroke-like episodes), MERRF (Myoclonus, epilepsy, and myopathy with ragged red fibers), NARP/MILS (Neurogenic muscular weakness, ataxia, retinitis pigmentosa/Maternally inherited Leigh syndrome), LHON (Lebers hereditary optic neuropathy) "Mitochondrial blindness", KSS (Kearns-Sayre Syndrome), PMPS (Pearson Marrow-Pancreas Syndrome), CPEO (Chronic progressive external opthalmoplegia), Leigh syndrome, Alpers syndrome, Multiple mitochondrial DNA (mtDNA) deletion syndromes, MtDNA depletion syndrome, Complex I deficiency, Complex II (SDH) deficiency, Complex III deficiency, Cytochrome C oxidase (COX, Complex IV) deficiency, Complex V deficiency, Adenine Nucleotide Translocator (ANT) deficiency, Pyruvate dehydrogenase (PDH) deficiency, Pyruvate carboxylase deficiency, Ethylmalonic aciduria with lactic acidemia, 3-Methyl glutaconic aciduria with lactic acidemia, Refractory epilepsy with declines during infection, Asperger syndrome with declines during infection, Autism with declines during infection, Attention deficit hyperactivity disorder (ADHD) with declines during infection, Cerebral palsy with declines during infection, Dyslexia with declines during infection, MNGIE (Mitochondrial myopathy, peripheral and autonomic neuropathy, gastrointestinal dysfunction, and epilepsy), MARIAHS syndrome (Mitochondrial ataxia, recurrent infections, aphasia, hypouricemia/hypomyelination, seizures, and dicarboxylic aciduria), ND6 dystonia, Cyclic vomiting syndrome with declines during infection, 3-Hydroxy isobutryic aciduria with lactic acidemia, Diabetes mellitus with lactic acidemia, Familial Bilateral Striatal Necrosis (FBSN), Aminoglycoside-associated deafness,

Dilated or hypertrophic cardiomyopathy, Wolfram syndrome and Renal Tubular Acidosis/Diabetes/Ataxia syndrome.

72. (New) The method according to claim 67, wherein the mitochondrial disorder is selected from the group consisting of MELAS (mitochondrial encephalomyopathy with lactic acidemia and stroke-like episodes), MERRF (myoclonus, epilepsy, and myopathy with ragged red fibers), NARP/MILS (neurogenic muscular weakness, ataxia, retinitis pigmentosa/maternally inherited Leigh syndrome), LHON (Lebers hereditary optic neuropathy, "mitochondrial blindness"), KSS (Kearns-Sayre Syndrome), PMPS (Pearson Marrow-Pancreas Syndrome), CPEO (chronic progressive external opthalmoplegia), Leigh syndrome, Alpers syndrome, multiple mtDNA deletion syndromes, mtDNA depletion syndromes, complex I deficiency, ND6 dystonia, complex II (SDH) deficiency, complex III deficiency, cytochrome C oxidase (COX, complex IV) deficiency, complex V deficiency, adenine nucleotide translocator (ANT) deficiency, pyruvate carboxylase deficiency, and pyruvate dehydrogenase (PDH) deficiency.

73. (New) The method according to claim 67, wherein said mitochondrial disorder is a secondary disorder caused by acquired somatic mutations, physiologic effects of drugs, viruses, or environmental toxins that inhibit mitochondrial function.

74. (New) The method according to claim 67, wherein the mitochondrial disorder is a deficiency of cardiolipin.

75. (New) The method according to claim 67, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

76. (New) The method according to claim 74, wherein the deficiency in a pyrimidine synthetic pathway is the uridine synthetic pathway.

77. (New) The method according to claim 74, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

78. (New) The method according to claim 77, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

79. (New) The method according to claim 67, wherein the mitochondrial disorder results in lower than normal uridine levels.

80. (New) The method according to claim 67, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

81. (New) The method according to claim 80, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

82. (New) The method according to claim 81, wherein the reverse transcriptase inhibitor is Azidothymidine (AZT), Stavudine (D4T), Zalcitabine (ddC), Didanosine (DDI) or Fluoroiodoarauracil (FIAU).

83. (New) The method according to claim 81, wherein the protease inhibitor is Ritonavir, Indinavir, Saquinavir or Nelfinavir.

84. (New) The method according to claim 81, wherein the DHOD inhibitor is Leflunomide or Brequinar.

85. (New) The method according to claim 67, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

86. (New) The method according to claim 85, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

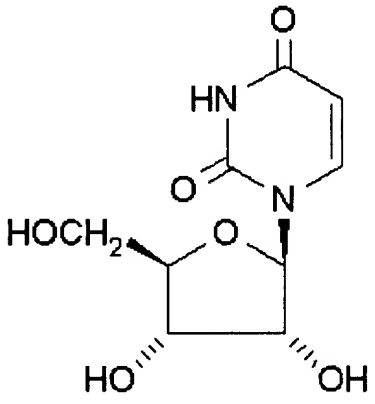
87. (New) The method according to claim 85, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

88. (New) The method according to claim 67, wherein the compound of Formula (I) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

89. (New) The method according to claim 67, wherein the compound of Formula(I) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

90. (New) The method according to claim 67, wherein the compound of Formula(I) is administered in a daily dosage of about 6.0 g/m^2.

91. (New) A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject in need thereof an effective amount of a compound of Formula I:



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92. (New) A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of uridine.

93. (New) A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of 1-b-D-ribofuranosyluracil.

94. (New) The method according to claim 91, wherein said symptoms are renal tubular acidosis (RTA), proteinuria, impaired eyesight, dementia, seizures, cardiomyopathy, skeletal myopathy, peripheral myopathy or autonomic myopathy.